



# Haemagazine

*For the Students, By the Students*

“To increase awareness of common hemato-oncological conditions.”

**Staff advisors:**

Dr. P. Ramachandran  
Dr. J. Julius Xavier Scott  
Dr. Leena Dennis Joseph  
Dr. Latha M. S.  
Dr. Dhaarani J.

**Editorial Board:**

Kapil Dev Nayar  
Shruti Iyer  
Chandni Agarwal  
Miloni More  
Pulkit Mehrotra  
Nuha Amrin Ali  
Sivaram T.

**Design Team:**

Wafiq Shimar Mohamed Jiyaudeen  
Miloni More  
Pulkit Mehrotra  
Nuha Amrin Ali  
Yashwanthy M. S.  
Arjun Suriakumar  
Veda Munnangi



# CHANCELLOR'S MESSAGE

**SHRI V.R.  
VENKATAACHALAM**

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I am glad to note that the M.B.B.S. students of SRMC & RI have brought out an e-magazine highlighting the social and health aspects of hemato-oncological problems in children.

I would like to congratulate the students and faculty who have made this possible.

Best wishes for continued efforts.

V.R. —



## VICE-CHANCELLOR'S MESSAGE

**DR. P.V.  
VIJAYARAGHAVAN**

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I am happy to note that the M.B.B.S. batch (interns) of SRIHER are bringing out a first-of-its-kind E-magazine titled "Haemagazine" which is focused on medical and social issues related to pediatric hemato-oncology. This will include topics of interest for exams in article format and imaging studies or lab investigations in quiz format.

The magazine is targeted not only at our institute students but also at alumni all over the world, and will be published on a quarterly basis and shared on social media.

The magazine is edited entirely by the interns editorial team, with staff advisors such as Dr. P. Ramachandran, Dr. J. Julius Scott and Dr. Leena Dennis guiding them.

I take this opportunity to appreciate the entire team for their stupendous effort.



## PRO-CHANCELLOR'S MESSAGE

### SHRI R.V. SENGUTUVAN

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Dear students,

It is a matter of great pleasure to know that the students of SRIHER are bringing out an E-magazine to create awareness, focusing on medical and social issues related to the field of pediatric hemato-oncology.

I wish to congratulate the editorial team of the magazine, pediatric hemato-oncology team and other senior professors of SRIHER guiding them, and wish them all success.

We, the administrators of SRIHER, will support them in all ways for improving the quality of care/life of these children. I feel proud that "future doctors" have shown this social welfare attitude at their young age, and I appreciate the motivation given by their mentors in the pediatric hemato-oncology department (Dr. Scott and his team).

All the best.

# DR. SCOTT'S MESSAGE

HOPE... MEDHOPE...

THE SQUIRREL - THE OFTEN QUOTED EXAMPLE OF THE EAGERNESS TO HELP OTHERS, WHICH REPORTEDLY HELPED LORD RAM BUILD A BRIDGE ACROSS THE SEA. THE ANECDOTE SIGNIFIES THAT EVEN SMALL ACTS OF KINDNESS AND HELP AT THE RIGHT TIME PLAY A HUGE ROLE.

THIS TINY RODENT THAT GOES AROUND CHEWING FRUIT DOES NOT KNOW THAT IT IS SOWING THE SEEDS OF TOMORROW'S TREES ALONG ITS WAY. THE SQUIRREL NEVER TAKES CREDIT FOR ITS ACT OF BENEVOLENCE TOWARDS ITS ENVIRONMENT (NOR DOES IT TAKE A PHOTO AND UPDATE ITS STATUS ON WHATSAPP...)

"SOW THE SEED, AND SOMEDAY IT SHALL BECOME A TREE. MULTITUDES SHALL BENEFIT FROM THE HARD WORK OF THEE"

STUDENTS ARE LIKE SQUIRRELS FOR "RAMA". THEY WILL HELP SOME "RAMA" NOW, AND THEY WILL BECOME THE FUTURE "RAMA" TO CREATE "RAMARAJIYAM."

I BELIEVE THAT...

WHEN WE INFLUENCE STUDENTS NOW THROUGH OUR VALUES, THEY WILL CHANGE OUR SOCIETY LATER. BUT WE SHOULD ALSO ACCEPT THAT A STUDENT'S LIFE IS TO BE ENJOYED AND WE SHOULDN'T FORGET THAT.  
AS PHILOSOPHER IMMANUEL KANT HAS SAID, "STUDENTS SHOULD ENJOY TO THE MAXIMUM WITH MINIMUM DISTURBANCE TO CO-HABITANTS", AND WE SHOULDN'T HINDER THAT.

ONE EXAMPLE OF THE IMPACT OF OUR SERVICE AMONG THE STUDENTS IS... A STUDENT AT OUR MEDICAL COLLEGE FOUND A 500RS NOTE LYING ON THE FLOOR AND WENT TO HAND IT OVER TO THE DEAN'S OFFICE, WITH THE REQUEST TO USE THIS MONEY TO TREAT A CHILD WITH CANCER IN G BLOCK.

I AM NOT RECOMMENDING THAT EVERYONE SHOULD CONTRIBUTE ONLY TO CANCER. THERE ARE SO MANY OTHER SPECIALITIES/NEEDY WHICH REQUIRE SOCIAL SUPPORT. WHAT I WANT TO EMPHASIZE IS THAT STUDENTS SHOULD HAVE SOME THINKING OF TAKING RESPONSIBILITY TOWARDS CONTRIBUTING TO THE SOCIETY FOR ITS WELFARE IN THE FUTURE.

AS ABDUL KALAM AND SWAMI VIVEKANANDA SAID, WE NEED THE YOUNGSTERS TO TRANSFORM THE SOCIETY.



OUR RACE IS "HUMAN"  
OUR RELIGION IS "LOVE"  
OUR LANGUAGE IS "KINDNESS"

YOURS,

A handwritten signature in black ink, appearing to read "J. Julius Xavier Scott".

DR. J. JULIUS XAVIER SCOTT  
MD, DCH, DNB (PEDIATRICS),  
FPHO (AUSTRALIA)

# *Tales of Tenacity:*

## Reborn in SRMC

In 2018, in a small village on the outskirts of Andhra Pradesh, a 3-month-old baby boy suddenly developed recurrent fever spikes and red patches all over his body. His father was barely able to make ends meet even for their household expenses, working as a driver, and his young mother had never travelled outside the boundaries of her village. Perplexed at the condition of their little one, this young couple rushed all the way to Chennai with hearts full of hope in search of a medical cure.

At a private hospital in Chennai, the doctors investigated the child extensively and then finally, the diagnosis of **Hemophagocytic Lymphohistiocytosis (HLH)** was made. Intensive chemotherapy followed by bone marrow transplantation (BMT) is the mainstay of treatment for this condition. The parents could make very little sense of these medical terminologies which were being hurled at them.

What started as a simple fever, came with a lethal diagnosis that required a BMT, leaving the parents absolutely crushed. There were several challenges ahead, such as finding a suitable donor for the Bone Marrow Transplant. Other challenges included the uncertainty of the outcome, the intercurrent infections and toxicities and, finally, the financial implications of all these. A BMT itself would cost 15 lakh Rs. and the pre- and post-transplant management (adding up to 25 lakh Rs.) only furthered this family's burden.

After having spent around Rs. 50,000 to arrive at the diagnosis which exhausted all of their savings, the family was shattered to realize that it was only the first step in a long, difficult and unpredictable journey. This disease, like any other hemato-oncological issue, sees no barrier and takes time for even an educated family to comprehend. With minimal education and facing a language barrier, this family found themselves in complete disarray. Above all, the chance of a successful outcome was only 50%, further highlighting the gap between effort and outcome in pediatric oncology.

After coming to SRMC, the child was admitted in the G Block ward. Our first ray of hope came when the sibling match was 10/10, which meant that the chances of a successful outcome were higher than usual. However, the mother's concerns multiplied, to think that her elder boy, the donor, would be subjected to several procedures. But the slightest hope kept their fighting spirit alive.

Hence began the tedious pre-transplant treatment to achieve remission. It was a particularly aggressive chemotherapy regimen, causing profound immunosuppression. The next 4 months of therapy were a battle between the medical team and the malady, to keep the HLH in remission. This period was interspersed with recurrent severely-debilitating infections, each of which served as a deterrent for us to proceed and would pull us right back to where we started. After 4 months of rigorous chemotherapy, the disease was finally in remission.

And now, the BMT could be implemented. BMT is an intense process starting with eradication of recipient stem cells followed by infusion of donor cells. Then comes the meticulous supportive care for the severely immunocompromised child until the donor stem cells start functioning, via a process called engraftment.

The BMT journey began with a neck vein being accessed, following which the child was conditioned with intensive drugs. To our dismay, however, the line was displaced. Many attempts to reinsert the line, by the most experienced team members, were unsuccessful. Hence, the child had to be shifted to the emergency OT for Infuse-a-Port insertion, an extremely high-risk procedure.

The next 24 hours were uncertain. The multi-disciplinary team, including intensivists, pediatric surgeons, nurses, doctors and anesthetists, fought tirelessly to keep the child alive until the stem cell infusion.

Just as things started looking better, the child developed a high-grade fever with an abysmally low WBC count (nearly zero) and had to be rushed into the intensive care unit. At this point all seemed lost to the young mother, who often forgot to eat or even drink water. The only solace the mother found was in her faith and her unconditional trust in the medical team. Just when the chances of her baby's recovery seemed bleak, nature answered her prayers and the bone marrow transplant finally seemed to kick in. However, with only partial uptake, the fear of subsequent rejection loomed. Foreseeing this upcoming turbulence, we modified the immunosuppressants and tackled all further infections aggressively.

India has always been enveloped by superstitions, and the mother was no exception to this. The unrelenting turmoil over the last 8 months had led her to believe that her child's misfortune had been her own doing. While the mother's pain was something we witnessed before our own eyes, the silent suffering of her husband and elder child was equally heart-wrenching. The father worked tirelessly at his minimum wage job and sent every penny towards his child's recovery, resulting in several sleepless nights where there was no food on his plate.

As for the elder child, separation from his mother at such a tender age for many months, was a struggle on its own. This ultimately led to the mother having an emotional breakdown, which needed intense psychological intervention. It was only after several sessions, where she was appreciated for her unwavering strength, that she began to recover.

But when the going gets tough, the tough gets going:  
***"Perseverance is not a long race; it is many short races, one after the other".***

And it was with this attitude that our team at SRMC embraced the mother and stood by her as her extended family. Determined to allay at least some of their many concerns, the whole of SRMC left no stone unturned in procuring 15 lakh Rs. for the child's treatment. From students to residents, nurses to doctors, social workers to NGOs, everyone came forward to ease their burden.

A lot of things were done to make this little boy's journey a success, but what deserves special mention is that no one chose to give up at any point. It's easy to be optimistic when things are going your way, but it needs real strength and resilience to remain hopeful in times of adversity. Though everyone played a small role individually, every drop makes an ocean and every brick makes a home. And each of us united to become the pillars of hope they needed.

Today, the child is growing and healthy. Though things are not always smooth sailing, every so often he shines his inquisitive look and graceful smile towards us. ***His zest for life continues to motivate us and emboldens our faith in what we as healers do.***



#### ***Hemophagocytic lymphohistiocytosis (HLH):***

- Uncontrolled activation of inflammatory cytokines.
- Accumulation of activated antigen processing cells (macrophages and lymphocytes).
- Presents with fever, hepatosplenomegaly, lymphadenopathy, cytopenias.
- Investigations reveal hypertriglyceridemia, hypofibrinogenemia, hyperferritinemia, low or absent NK cell activity, elevated soluble CD25 levels, with hemophagocytosis in lymph nodes, bone marrow, and spleen.
- Can be primary (familial) or secondary. Primary is caused by genetic mutation and secondary is associated with immunodeficiency states, infections, and auto-immune disorders.
- Early diagnosis is essential, as diagnostic delay has poor prognosis.

#### **Dr. Scott's Message:**

*When I think about this case, what strikes me is how Parthasarathy was with us from January to October, an extremely difficult ten months, where he was reborn in the womb of SRMC. I would like to extend my heartfelt thanks to the incredible team that joined hands to save the boy. Special thanks to PICU team under Dr. Shuba (H.O.D. Pediatrics), Dr. Prakash Agarwal (Head of Pediatric Surgery) and his department, Dr. Krishnamoorthy and the Department of Transfusion Medicine, Dr. Ranjith and his team in the Department of Cardiac Anesthesia and all the duty doctors, nurses, pediatric residents, social workers and administrative officers for their contribution. Lastly, it gives me immense pleasure to express my appreciation to one of our own, Dr. Dhaarani, a young pediatric BMT physician. She took the challenge head-on in the early years of her career and was instrumental in the child's recovery. My sincere thanks to our Founder Chancellor, Chancellor, Pro-Chancellor, Vice-Chancellor, Deans, medical director, medical superintendent, nursing superintendent, all administrators and the whole of the SRMC family for the opportunity to help this child.*

# *Tales of Tenacity:*

# Know the Glow

**This story tells us the importance of systematic examination of every infant and how a simple red eye reflex examination would have changed the life of this little girl:**

In a village in North India, there lived a farmer Ramesh and his devoted wife, Ganga. The two were always content with their simple lives, but for many years they had yearned for a child. Five years passed by, and just as they were losing hope, they were finally blessed with a baby girl. Being the only child of her doting parents, she was the apple of their eyes. Relatives from near and far came to visit the beautiful baby. Each of them was smitten immediately by the twinkle in her eyes.

A year flew by and the child grew healthy as ever. But soon, they began to grow suspicious of the white gleam in her eye. Concerned, they took her to the nearby Primary Health Centre where she was given nutritional supplements. The parents meticulously administered her vitamins every day hoping for some change, but, much to their confusion, the whitish glow remained. After 8 months of fruitlessly continuing the treatment, they sought a nearby Ophthalmology Centre. She was then diagnosed with traumatic cataract, which initially frightened the parents, but they were reassured that a simple surgery would allay her problems. The surgery was successful and she was on regular follow-up. The parents breathed a sigh of relief.

Just when the family thought that they had put the worst behind them, they began to notice a swelling in the left eye. As it grew day by day, so did their concerns. They rushed her to the Ophthalmology Centre and imaging revealed **Metastatic Retinoblastoma**. The parents could not believe what they had been told. The very twinkle that they had once adored, had reared its ugly head. They were advised intensive chemotherapy, which left them shattered. Finding themselves in the midst of the COVID pandemic, help was extremely hard to get. As they scrambled, gathering every penny to organize funds for the chemotherapy, the swelling continued to grow. It had now managed to envelop the entire left half of her face, ulcerating in multiple places. This giant growth had disfigured her once innocent, smiling face. The foul-smelling discharge emanating from the growth, increased the urgency. After tremendous efforts, she was finally brought to Chennai to Sankar Nethralaya Eye Hospital and she was referred to SRMC for chemotherapy. The MRI showed extensive brain metastasis, posing a terminal diagnosis for the child. On hearing this, the parents were distraught. Their whole world came crumbling down. The pain that their little one had to endure crushed their spirit.

When the diagnosis is so grave, the next step of treatment is often a challenge. Upon discussion with the tumor board, we planned for palliative chemotherapy and supportive care, the costs of which were borne by our hospital and NGO (Tiara Hemophilia and Cancer Foundation) in view of their financial concerns.

After the first cycle of chemotherapy itself there was significant reduction in the swelling of her eye. She was visibly better, her appetite had improved and she showed glimpses of the lively child she had once been. The family was incredibly grateful for these last few months with their little angel. Unfortunately, death was inevitable. After 4 cycles of intense chemotherapy, the disease had continued to progress to her brain and she succumbed to her illness.

This case has left a deep impression in our hearts and its recollection reminds us that we are not invincible. When a case presents in the final stages, there is often little we can do beyond making their last days comfortable.

While we have witnessed several cases of retinoblastoma, this case made us realize the massive impact of early screening and intervention. Retinoblastoma, a common intraocular malignancy is one of the most curable cancers in children. Therefore, success stories lie in early detection, timely diagnosis and optimal treatment. Lack of awareness among general public and primary care physicians about the 'white eye reflex' led to missed opportunities of identifying the disease at an earlier stage in this child. Most of the pediatric malignancies cannot be prevented or identified early by screening. Retinoblastoma is the only pediatric cancer that can be identified at a very early stage. Every new born and young children admitted to a hospital should therefore be periodically screened for the red eye reflex. The best treatment for retinoblastoma lies in raising public awareness about the '**White Eye Reflex**'.

**Hence lets all vow to "Know the Glow" and remember that red is not always a danger sign.**





## **Red Eye Reflex:**

*The red reflex is a reflective phenomenon seen when light passes through the pupil and is reflected back off the retina to a viewing aperture, creating a reddish orange glow.*

*Red reflex testing is a valuable tool for detection of abnormalities in the normally transparent visual axis or in the retina.*

## **White Eye Reflex (Leukocoria):**

*In contrast to the normal red reflex, leukocoria is defined as a yellow, pale, white, or otherwise abnormal reflection of light observed in the pupil. It is the earliest sign of retinoblastoma in children.*

*The American Academy of Pediatrics currently recommends red reflex assessment as a component of the eye evaluation in the neonatal period and during all subsequent routine health visits in children less than 5 years of age.*



## **Dr. Scott's Message:**

*Apart from educating ourselves about white eye reflex, after watching the child's quality of life improve with palliative intent chemotherapy and supportive care and although we couldn't cure the child, it reminded me of one of the teachings of the American Academy of Pediatrics: "**Palliative care is not adding years to life, but adding life to years**".*

**"Cure sometimes, relieve often, comfort always"**, these great words were said by none other than Hippocrates himself and we find how deeply it needed to be enforced in this case.

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U  
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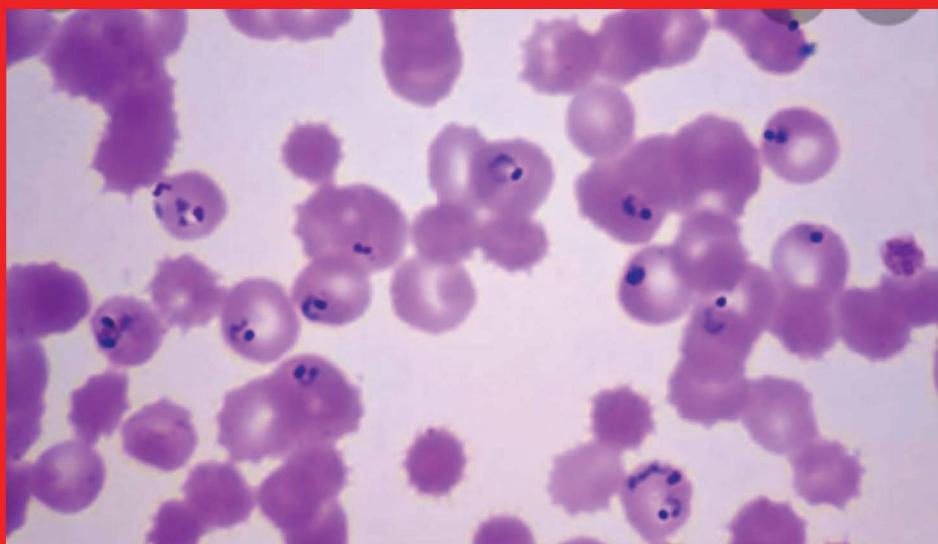
## Question #1

Where was this "bubble"-ing child living and what was the name of the disease he was suffering from?



## Question #2

- A. Identify the striking abnormality in the cells.
- B. What is the drug used for the treatment of this disease that can cause hemolysis?



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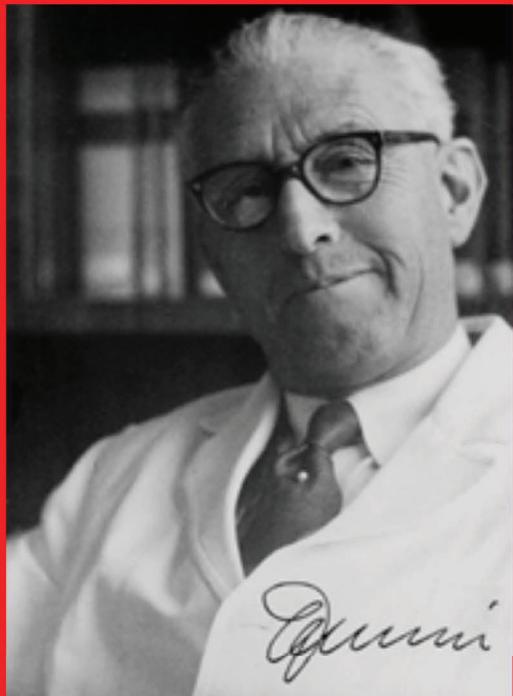
## Question #3

A 10 year old child presented with pancytopenia and short stature.

Given below is the famous Swiss paediatrician who discovered the disease she has.

A. Identify the Doctor.

B. Spot the diagnosis.



# Human Papilloma Virus Awareness

Do you know enough about a jab that can prevent cancer?



## Vaccine preventable cancer

Cancer of the cervix is the only cancer that is almost entirely preventable through a safe, effective vaccine and screening protocol.

About 96,922 new cervical cancer cases are diagnosed annually in India (estimates for 2018). Cervical cancer ranks as the 2nd leading cause of female cancer in women aged 15 to 44 years in India.



Did you know?

4TH

MOST COMMON CANCER IN WOMEN



7TH

WORLDWIDE



2ND

IN INDIA

## HPV VACCINATION IN INDIA (IAP RECOMMENDATION)

- Minimum age: 9 years
- HPV4 [Gardasil®] and HPV2 [Cervarix®] are licensed and available
- Only 2 doses of either of the two HPV vaccines (HPV4 & HPV2) for adolescent/preadolescent girls aged 9-14 years, the doses are administered in 0-6 months schedule.
- For girls 15 years and older, and immunocompromised individuals 3 doses are recommended. The schedule is HPV4 (0, 2, 6 months) or HPV2 (0, 1, 6 months)
- For two-dose schedule, the minimum interval between doses should be 5 months.



Great! When should I get vaccinated?

WHO recommends HPV vaccination for girls alongside screening and treatment for older women to reduce their cancer risks.

The vaccine is most effective when given early in adolescence between the age of 9 and 14 years - BEST before girls are exposed to the virus.

So, is this a worry only for females?

Nope! There is growing evidence of HPV being a relevant factor in other anogenital cancers (anus, vulva, vagina, and penis) as well as head and neck cancers.

What test is used for screening?

A liquid-based technology called the Papaincolaou smear, taken at the transitional zone of the cervix - between the endocervix and ectocervix.



# The MedHOPE Foundation

A foundation by Medical Students for  
Hemato-oncology patients

**MedHOPE** is a students' organization started in 2011 at **Sri Ramachandra Institute of Higher Education and Research** by MBBS students, in order to help cancer-afflicted children in a holistic manner.

MedHOPE won the **Most Innovative Project of Cancer Support** award in 2013 at National Level in Tata Memorial Hospital, Mumbai.

MedHOPE conducts Rose Day, International Childhood Cancer Awareness Day, blood donation camps, walkathons, play day activities and is actively involved in organizing fundraising events for children with cancer.

They also conduct an annual MBBS undergraduate exam refresher course – **HOPE**, wherein apart from the final year exam oriented-training, MBBS students are also exposed to career options discussions. The funds generated through this conference are donated towards aiding the treatment of cancer-afflicted children.



Commemorating the **Rose Day on September 22nd**, this year the MedHOPE volunteers donated roses and gifts to the children with cancer staying in the ward. They also organized a **blood donation drive** from 22nd to 24th September.

Blood Donation Camp 2018

## PATTIMANDRAM

கலைப்பு: சமூக வளைவாளர்கள் / தொலைதொடர்பு சாலை

Social networking / Mobile communication

திருச்சியில் நடைவாழ்வு மற்றும் நிதி வாழ்வு

Solomon Pappaiah Pattimandram 2017

HOPE Conference 2016



Chinmayi Live-In Concert 2016

S. P. Balasubrahmanyam Concert 2014

# THCF – The Tiara for Our Cancer Children

**Tiara Haemophilia and Cancer Foundation** is an NGO that supports the treatment of poor children with cancer and haemophilia. It is due to the financial support of THCF that so many lower economic status families have been able to initiate cancer treatment for their children. As a result, the treatment refusal rate or treatment non-compliance has been less than 1% in our centre.

**Aparna Guhan Shyam**, the **founder of THCF** is an epitome of benevolence, authenticity and impeccability. Let's hear what she has to say about herself and her brainchild NGO:

## 1. Tell us about you and your background. How did it all start? Was there any inspiration/incident/mentor who motivated it?

I learnt briefly about Haemophilia when I was in 4th standard in school. It stayed in my mind over the years. When I was 14 years old, I decided I had to do something to help, so I started donating my pocket money and money from birthdays and festivals to help patients with Haemophilia. By the time I was 16, opening a charity became my dream. I even had the name Tiara picked out. I donated like this for 10 years. At the age of 24, I came back from London after finishing my Masters and within a month of being back, I started Tiara Haemophilia and Cancer Foundation (THCF).

## 2. Can you brief us about all the activities in your foundation?

Tiara Haemophilia and Cancer Foundation (THCF) is a Children's Charity that supports children from low socio-economic backgrounds who have been diagnosed with Haemophilia or Cancer. Our support is both financial and emotional, providing these children and their families with a support system and helping them take a vital step towards a better quality of life.

We also spread awareness about Haemophilia and Childhood Cancer and support medical research in these fields, with a view to improve the lives of affected children and their families.

## 3. How far do you feel your dream has been achieved till now?

When I started THCF, I never expected it to become so big. It was started with just a passion to help people. We signed an MOU with SRMC the first year and started helping the patients in G Block. Over the years, we expanded our network. Our support is currently available in 14 hospitals across Chennai, Hyderabad, Coimbatore and Madurai and the list keeps growing.

We've enabled over 500 children from low socio-economic backgrounds to access quality Haemophilia and Cancer care. I'd say we've more than achieved the initial dream.

## 4. What is your ultimate dream?

The scope is being enhanced as the years go by so the dream keeps growing. The minute we define our ultimate dream, we limit ourselves. We look forward to what is to come.

## 5. Is family life interfering with your work in THCF or vice versa?

No. My family is very supportive of my work.



and we've just begun.

## **6. Can you tell us about the awards & recognitions that your foundation has got so far?**

- Sri Ramachandra University honored me for making a difference in the lives of children with Cancer and Haemophilia and their families.
- Honoured us for our work at International Childhood Cancer Day celebrations at Sri Ramachandra Hospital.
- Honoured at the Childhood Cancer Survivors Day Celebration for being the major contributor to Bishal Baghat's Bone Marrow Transplant, the first such transplant at Sri Ramachandra Hospital.
- Dr. Muthiah's Virtues Trust & Dr. Visalakshi Muthiah Foundation for Special Children honoured us for our immense and untiring crusade against Haemophilia and Cancer in children.
- Honoured by the Haemophilia Society - Tirunelveli Chapter for our continuous love and support in caring towards children with Haemophilia on the occasion of their Founder's Day Celebration, Disability Card Distribution & Annual Year Celebration.
- Honoured by Le Midaz International & Susrutha Medical Research Foundation for the valuable contribution and outstanding performance in the field of health

## **7. Any hurdles/difficulties/insults faced so far in your journey?**

Yes, we've faced several challenges over the years and will continue to do so but we always overcome them.

## **8. How do you think students at SRMC can help you in your endeavour?**

We would love SRMC students to volunteer for our fundraising events.

## **9. Was there any incident/family that touched you?**

We've formed bonds with so many families over the years. They are all special and that's what makes this so rewarding. The sheer strength we see from these children and their parents is heartening.

## **10. Do you want to acknowledge anyone for your success?**

We have a great Board of Trustees and Medical Advisory Board, a hardworking and passionate team, and strong partnerships with donors, hospitals, distributors and doctors. All these contribute to our success.

## **11. Any advice for SRMC students/medical students?**

Medical students, you can create a lot of public awareness about Haemophilia and Childhood Cancer.



**Aparna Guhan Shyam**  
*Founder of THCF*

# A N S W E R S

## Question #1

The photo shows David Vetter, a child suffering from **Severe Combined Immunodeficiency (SCID)**.

It is also known as the “**bubble baby disease**”. Children affected by SCID are extremely vulnerable to infectious diseases and hence, before the era of antibiotics/BMT, some of them would live in a sterile environment - like the bubble shown in the photo.

### Salient features of SCID:

- Primary immunodeficiency disease with profound T-lymphocyte defect.
- It usually manifests **within 6 months of life** and is considered as a **MEDICAL EMERGENCY**.
- It is an emergency indication for **BONE MARROW TRANSPLANT** (only curative option).
- **EARLY DIAGNOSIS** is the key to effective management.
- Comprehensive family history is important – e.g. early sibling deaths, recurrent infections, consanguinity.
- Pointers towards SCID - absolute lymphocyte count **ALC <3500/cubic mm in INFANCY** or **ALC ≤1500/cubic mm beyond INFANCY**.
- **TREC – T-cell excision circles** – by-product of T-cell production used as a **NEWBORN SCREENING METHOD** for early diagnosis.

# A N S W E R S

## Question #2

- A. Malarial parasites within RBCs - ring stage.
- B. Primaquine.

In cases of high parasite load, it can cause a severe acute intravascular hemolytic episode leading to circulatory failure, severe anemia and renal failure - called **BLACKWATER FEVER**.

The drug primaquine can also cause an intravascular hemolysis in children who have **G6PD deficiency**.

# A N S W E R S

## Question #3

A. Dr. Guido Fanconi

B. Fanconi anaemia

**Guido Fanconi**, a Swiss paediatrician, was regarded as one of the founders of modern paediatrics. There are several medical conditions named after him. In 1927, he described hereditary panmyelopathy with short stature and hyperpigmentation, known as Fanconi anaemia. In 1934, the first cases of cystic fibrosis of the pancreas were described in a thesis written under his direction. His contribution to renal physiology also lead to his name being lent to Fanconi syndrome.

### Salient features of Fanconi Anemia:

- **Autosomal recessive disorder with pancytopenia**, which usually manifests at the end of the 1st or 2nd decade of life.
- **Thumb anomalies** (bifid thumb, hypoplastic thenar eminence), dysmorphism, short stature, Café-au-Lait macules, renal anomalies (usually ectopic kidney) and genitourinary anomalies are characteristic features.
- Being a premalignant condition, these patients are prone to **develop hematological malignancies**, especially Acute Myeloid Leukemia (AML) and squamous cell carcinoma of the head and neck.
- **Bone marrow transplant** corrects the hematological manifestations and improves the quality of life.



# Haemagazine

*For the Students, By the Students*

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